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LIVING WITH PULMONARY FIBROSIS

Contents

In this booklet, you can find out about the different types of pulmonary fibrosis in adults, common symptoms, and how it's diagnosed and treated.

Pulmonary fibrosis is scarring of the lungs. It's the end result of many conditions, called interstitial lung diseases (ILDs). There is no cure for pulmonary fibrosis, but the right care can help you feel better and improve your quality of life. We've put this information together for you, your family, carers, and friends.

pulmonary = it affects your lungs

fibrosis = the name for a build-up of scar tissue, which makes your lungs stiff. You might hear 'fibrosis' used to mean hardening

What is pulmonary fibrosis?

Pulmonary fibrosis is the buildup of scar tissue in your lungs. It makes breathing increasingly difficult.

Some types of pulmonary fibrosis have an identifiable cause. But for many types, a definite cause cannot be found.

Although we do not always know what causes pulmonary fibrosis, we do know it is not a form of cancer or cystic fibrosis, and it's not contagious.

What are interstitial lung diseases?

Interstitial lung disease (ILD) is the name for a group of lung conditions that damage the lung tissue. There are more than 200 different ILDs.

Interstitial lung diseases cause scarring in your lungs (pulmonary fibrosis), inflammation in your lungs or a mix of both. Some mostly cause scarring. Some mostly cause inflammation. Often there is a combination of scarring and inflammation. It's important to know what the major cause of your symptoms is to help your health care team decide the best treatment for you.

interstitial: affects your 'interstitium', the network of tissue that supports the air sacs in your lungs

Our information covers the ILDs seen most often:

- IPF
- Sarcoidosis
- Hypersensitivity pneumonitis
- Occupational interstitial lung diseases
- Connective tissue and autoimmune diseases
- Drug-induced ILDs

What's the outlook?

Most forms of pulmonary fibrosis tend to get worse over time. The likelihood of this happening and how quickly it happens varies with the type of pulmonary fibrosis, but also from person to person. This makes it difficult to predict exactly how fast symptoms may progress and change. The aim of treatment is to slow down this rate of change. The chance of success depends on the exact diagnosis and how well you can tolerate treatment. You will be monitored and have regular tests of your lung function to look for changes.

In some people, pulmonary fibrosis progresses very quickly, while for others their condition remains stable for many years. Some conditions progress at a steady rate. In others, apparently stable conditions can change suddenly. This sudden worsening of symptoms is called an exacerbation.

There are no defined stages in pulmonary fibrosis, but doctors may talk about mild, moderate, severe, or end-stage fibrosis.

Questions about diagnosis, treatment and outlook?

The treatment and outlook for different types of pulmonary fibrosis varies considerably. If you're not sure about your diagnosis, check with your doctor or nurse for the exact name of your lung condition.

If you are unsure what to ask your health care professional when you have, or could have, pulmonary fibrosis, read our helpful list at **blf.org.uk/support-for-you/pulmonary-fibrosis/questions-to-ask**. It suggests questions to ask about diagnosis, treatments and much more.

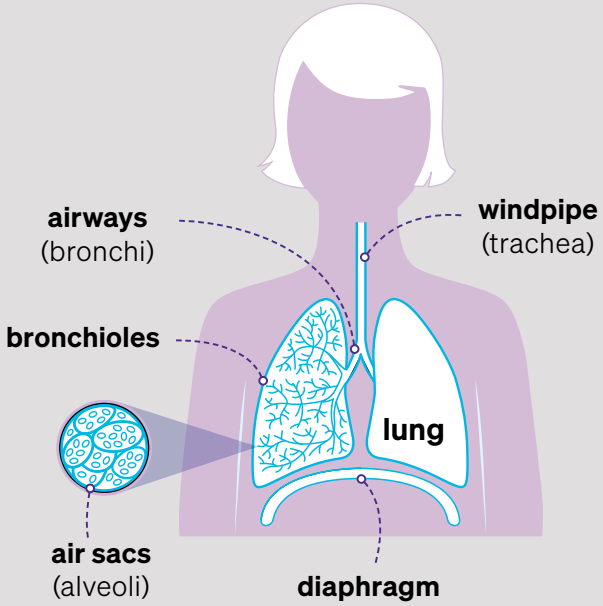
How does pulmonary fibrosis affect your breathing?

All kinds of pulmonary fibrosis cause scarring in your lungs that make breathing more difficult. Scarring makes your lungs stiffer and less elastic so they're less able to move and take oxygen from the air you breathe.

How do you breathe?

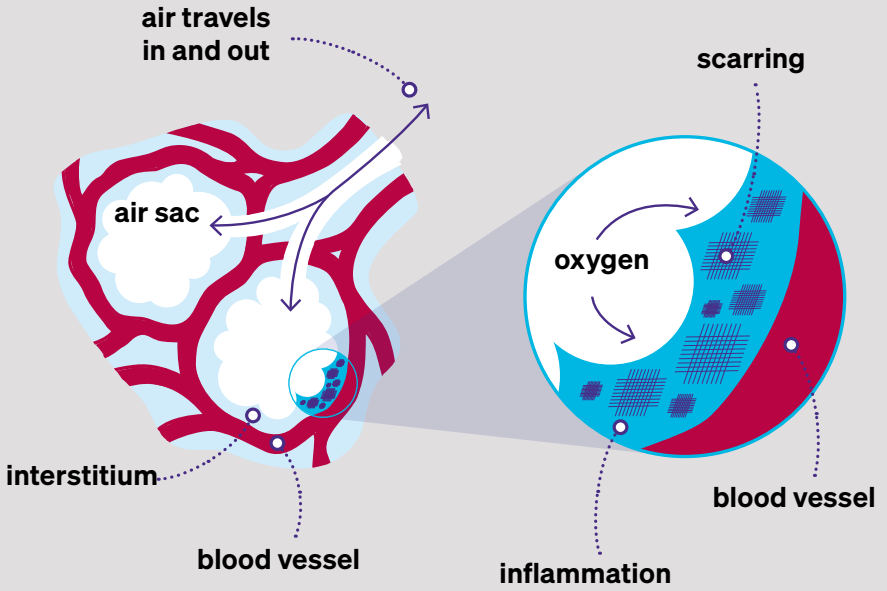
Each time you breathe in, you draw air into your nose or mouth, down through your throat and into your windpipe. Your windpipe splits into two smaller air tubes called bronchi, which go to your lungs. The air passes down the bronchi, which divide again and again, into thousands of smaller airways called bronchioles.

The bronchioles have many small air sacs, called alveoli. Inside the air sacs, oxygen moves across paper-thin walls to the capillaries – tiny blood vessels – and into your blood. The air sacs also pick up the waste gas, carbon dioxide from your blood, ready for you to breathe it out.



How does pulmonary fibrosis change your breathing?

If you have pulmonary fibrosis, scarring affects the air sacs in your lungs. The air sacs are supported by the 'interstitium', a network of supporting tissue. Scarring happens in the gaps between and around the air sacs and limits the amount of oxygen that gets into the blood.



As scarring increases, your lungs are less able to expand to allow you to take deep breaths and the level of oxygen in your blood can start to drop. Breathing may feel like harder work and you can feel breathless from everyday activities like walking.

Types of pulmonary fibrosis

Idiopathic pulmonary fibrosis (IPF)

Idiopathic pulmonary fibrosis (IPF) is a lung condition that scars your lungs and makes it difficult for you to breathe. It's the most common type of pulmonary fibrosis.

Idiopathic means the cause is not known, but researchers now believe that the body creates fibrosis (scarring) in response to an injury to the lung. The initial injury to the lungs might be from:

- **acid reflux** from the stomach
- **viruses** – in some studies, IPF has been linked to certain viruses, including herpes, hepatitis C and the Epstein Barr virus, which causes glandular fever
- **environmental factors** – breathing in certain kinds of dust from wood, metal, textiles or stone, or from cattle or farming
- **cigarette smoke** exposure.

It is not known if these factors directly cause IPF.

Some people may also get IPF if it's in their family history. But this link is only found in a minority of cases. According to a 2021 survey by the British Thoracic Society, 5.7% of patients were found to have a known relative with IPF.

What are the symptoms of IPF?

If you have IPF you may feel short of breath a lot, and not just when you're moving about. If you feel breathless you should visit your GP. A cough that doesn't go away (usually a dry cough) and feeling tired are other symptoms of IPF.

You may also notice clubbing of your fingertips or toenails. This means the shape of your fingers or toes might appear like a drumstick, with the tips being larger around the nails. The base of the nails may also feel spongy. Having this doesn't mean you have IPF as it can occur in other conditions too. But check with your GP if you notice it developing.

How is IPF treated?

Medication to slow the scarring

There are currently two drugs that can be prescribed to slow down the rate of scar tissue developing in the lungs:

- Pirfenidone
- Nintedanib

These drugs can only be prescribed to people whose lung function is within a set range. If your lung function is outside of that range, you might only be able to access treatment by taking part in clinical trials.

If you're eligible, your specialist team will decide which drug is best for you. Current studies suggest both drugs are equally effective in slowing down the rate of scarring in the lungs.

Pirfenidone – brand name Esbriet

This treatment comes in the form of capsules or tablets taken with meals. Clinical trials found that the drug slowed down the loss of lung function in most people with IPF, slowed the rate at which their symptoms got worse, and improved life expectancy.

Some people also might have skin reactions to sunlight. Talk to your doctor about possible side effects if you're considering taking Pirfenidone and ask about using sunscreen to prevent skin problems.

Nintedanib – brand name Ofev
Nintedanib has also been shown in trials to slow the rate at which lungs become scarred in IPF. This drug is also taken in the form of capsules.

If you're taking certain medications, such as blood thinners, you may be told not to take Nintedanib.

Before the availability of specific treatments, studies showed that almost 5 in 10 people with IPF in the UK died within three years of their diagnosis. But some people, about 2 in 10, lived for more than five years after they were diagnosed. Clinicians now believe the treatments available will mean that people diagnosed today will live longer.

Find out more about IPF at **blf.org.uk/IPF**

Sarcoidosis

Sarcoidosis – also called sarcoid – is a condition where inflamed cells clump together to make small lumps called granulomas. These granulomas can develop in any part of your body.

They are most commonly found in the lungs and the lymph glands which drain the lungs. They can also affect your skin, eyes, joints, heart, nervous system, liver, spleen, muscles, nose and sinuses.

When lots of granulomas develop in one area, they begin to affect how well that part of your body works. This causes symptoms. The inflammation, which is usually reversible, can sometimes progress to scarring, which isn't reversible.

What are the symptoms of sarcoidosis?

Symptoms of sarcoidosis include:

- feeling short of breath
- a cough which is often dry
- tiredness
- feeling ill or feverish
- red, painful eyes with impaired eyesight
- painful red lumps on your shins
- swollen glands in your face, neck, armpits or groin
- skin rashes
- painful joints, bones or muscles
- an abnormal heart rhythm or chest pain
- sweats
- fatigue that, when severe, can be extreme.

How is sarcoidosis treated?

Sarcoidosis can get better without medication. This means it's normal for your doctor to keep an eye on your symptoms for a few months before talking about treatment. It's usual to have regular chest X-rays, breathing tests and blood tests to monitor your condition.

Many people with sarcoidosis don't need treatment.

But treatment may be needed if:

- symptoms are affecting your quality of life
- scarring in your organs is severe enough to be potentially dangerous.

Most people with acute sarcoidosis, which is short term, won't need specific treatment. If your sarcoidosis is causing you pain, such as muscle or joint pain, a painkiller such as ibuprofen or paracetamol can help.

Treatment for sarcoidosis is aimed at **improving your symptoms and preventing inflammation from causing scarring and damage** to the affected parts of your body.

Steroids

Steroids are an effective treatment. Sometimes they can be used directly on the part of your body that's affected. For example, you can use eye drops for eye symptoms. More often, you'll take them as a course of tablets.

If you take high doses of steroids for a long time, you can experience side effects. These can include increased appetite and weight gain, indigestion, heartburn, mood disturbance and difficulty sleeping. They can also cause thinning of the bones, or osteoporosis.

For this reason, you'll usually take a high dose of tablets for a short time, followed by a lower dose over a much longer period. If you stop taking steroids too soon, your condition might become active again and cause more scarring. So, you'll often need to continue the treatment for up to two years.

Often you'll only need one course of steroids, but sometimes you might need to take a second course. Only a small number of people with sarcoidosis need long-term treatment with steroids. If you take steroids long term, most health care centres will recommend a bone density scan. This may be repeated if you take steroids for over 2 years.

Other medication

For a very small number of people, steroids are not enough to control their symptoms, or the dose needed to control symptoms in the longer term is too high. These people may need to take other medications called immunosuppressants that help control your body's immune system, such as methotrexate, azathioprine, hydroxychloroquine and mycophenolate mofetil (MMF).

These all require careful monitoring with regular blood tests and may increase your risk of infection. Some have side effects that you should discuss with your health care professional.

Hypersensitivity pneumonitis

Hypersensitivity pneumonitis (HP) happens if your lungs develop an immune response – hypersensitivity – to something you breathe in which results in inflammation of the lung tissue – pneumonitis. It used to be called extrinsic allergic alveolitis (EAA).

One example is **farmer's lung**. This is caused by breathing in mould that grows on hay, straw and grain. Another is **bird fancier's lung**, caused by breathing in particles from feathers or bird droppings. It can be very difficult to find the exact cause of hypersensitivity pneumonitis.

What are the symptoms of hypersensitivity pneumonitis?

The symptoms, including coughing, shortness of breath, and sometimes fever and joint pains can come on suddenly after you've been exposed. This is the acute (short-term) form of the condition. It can go away – without leading to fibrosis (scarring) of the lung – if you can permanently avoid the substance that caused the attack.

In other cases, symptoms of breathlessness and cough may only appear more gradually, perhaps over many years, as a result of permanent scarring of the lungs. This is called chronic (long-term) hypersensitivity pneumonitis and often a specific cause cannot be found.

How is hypersensitivity pneumonitis treated?

Hypersensitivity pneumonitis is regarded as a more treatable cause of pulmonary fibrosis, but it can cause symptoms that get worse over time and become hard to treat.

If a specific cause is identified, it's **really important to completely avoid exposure to it**.

You may need to take anti-inflammatory medication called steroids for a few weeks or months. If you need steroids to control the condition for longer, your doctor may recommend more drugs to reduce the risk of side effects associated with steroids.

"I may never know what's causing my condition."

Jane, 61, was first diagnosed with hypersensitivity pneumonitis 10 years ago

When I moved to London, I developed a cough. It got so bad I ended up in hospital and was diagnosed with hypersensitivity pneumonitis. I took high dose steroids – and my symptoms disappeared! For seven years I had no symptoms at all.

But when the symptoms came back, they didn't go away. So now my hypersensitivity pneumonitis is long-term. Tests showed I have a hypersensitivity to pigeon and budgie droppings, but my doctor says there are thousands of other things that I could be reacting to. I may never know what's causing my condition.

I get unpleasant bouts of coughing. And small things make me tired – like carrying shopping home.

I found my work as a lawyer more and more difficult. My employers suggested I claim on their permanent health insurance policy. My claim was accepted and I'm now on long-term sick leave.

I take steroids and immune-suppressing drugs every day. I'll probably take drugs for the rest of my life.

Occupational interstitial lung diseases

Pneumoconiosis is a term for a group of lung diseases caused by breathing in specific dusts in your workplace. They get lodged inside your lungs and cause scarring.

The most common type is coal worker's pneumoconiosis, caused by breathing in coal dust. Other forms are silicosis, caused by exposure to silica dust, and asbestosis, caused by breathing in asbestos. Although they are types of pneumoconiosis, silicosis and asbestosis are treated as separate pulmonary fibrosis conditions.

There is often a long delay (10 years or more) between breathing in the dust and showing symptoms, so new diagnoses usually reflect past working conditions. People are often retired by the time they're diagnosed.

If you've been exposed to certain substances in the course of your work, you may be entitled to compensation or benefits.

What are the symptoms of pneumoconiosis?

Symptoms of pneumoconiosis are similar to other forms of pulmonary fibrosis, including coughing, feeling breathless or feeling very tired. People with coal workers' pneumoconiosis may cough up black mucus (phlegm).

How is pneumoconiosis treated?

The main treatment is avoiding the dust or fumes causing the condition. There are no specific drug treatments. Oxygen therapy and pulmonary rehabilitation may help with your symptoms.

Connective tissue and autoimmune diseases

For reasons we don't fully understand, sometimes the immune system turns against the body. This is known as autoimmune disease. When your immune system attacks your body's own connective tissues, they cause scarring (fibrosis).

Connective tissues lie under the surface of your skin and around your internal organs and blood vessels. If an autoimmune disease, including rheumatoid arthritis, Sjögrens syndrome and systemic sclerosis (scleroderma), affects your lungs, they can cause pulmonary fibrosis (scarring of the lungs). This is sometimes called a connective tissue disease-related ILD or CT-ILD.

Unfortunately, some of the drugs used to treat these autoimmune diseases may rarely also cause lung damage as a side effect.

The tendency to develop some forms of auto-immune or connective tissue diseases is genetic. If you have this type of pulmonary fibrosis, your doctor should ask about your family history of these as well as other lung diseases.

What's the outlook of pulmonary fibrosis caused by autoimmune disease?

Your prognosis (likely course of your condition) will depend on many factors, including the particular form of autoimmune disease you have, how severe it is and the way it affects your lungs. Some people live just a few years after their diagnosis, particularly if they develop complications such as pulmonary hypertension. But other people live much longer. Talk to your doctor about your individual situation.

What's the treatment for pulmonary fibrosis caused by autoimmune disease?

You might need to be under the care of both a rheumatologist and a respiratory specialist. You'll usually be treated with immunosuppressant drugs.

An antifibrotic drug – called Nintedanib – has also been shown to help slow the progression of pulmonary fibrosis, in people where the fibrosis is progressing despite immunosuppressant drugs. Nintedanib might be recommended as well as immunosuppressant drugs, or sometimes instead of, depending on the individual.

As well as treating lung symptoms, managing your underlying condition is essential to protect your lungs from more damage.

Help to stop smoking

If you smoke, stopping is very important for your lung condition and overall health. Your GP can refer you to free help to stop smoking.

Drug-induced interstitial lung disease

Any medication can have side effects. In rare cases, medicines can damage the lung tissue and cause inflammation which may cause pulmonary fibrosis (scarring of the lungs).

Some of the more common medication types that are known to carry the risk of pulmonary fibrosis include certain:

- antibiotics, particularly nitrofurantoin
- immunosuppressant drugs, such as methotrexate
- drugs for heart conditions, particularly amiodarone
- cancer chemotherapy drugs
- biological agents used to treat cancer or immune disorders.

There are many other drugs that can potentially cause ILDs. Before starting any medication, you and your doctor will weigh up the risks and benefits. Sometimes the choices are difficult, especially if it's a life-saving treatment. It's important to tell your doctor about any new symptoms you have.

The situation varies for each individual and for each drug. Breathing problems from drug-induced pulmonary fibrosis can come on suddenly or develop more slowly over time.

What's the treatment for drug-induced interstitial lung disease?

If it's found a drug you're taking or treatment you're on is causing interstitial lung disease, you'll be advised to stop the treatment to prevent any further damage, and your treatment will be changed in consultation with your specialist or GP. You might also be prescribed immunosuppressant treatment (such as steroids) to help your body's immune response.

What's the outlook for drug-induced interstitial lung disease?

If a drug has caused fibrosis, people often get better quickly if the medication is stopped before much damage is done. Identifying this problem and stopping the drug is the key intervention.

Steroid medication can help calm down your body's response to the medication. But some people may have lasting lung damage. The prognosis (likely course of the condition) depends on the medication that caused the fibrosis, how much fibrosis occurred and the person's overall health. Most forms of drug related ILD aren't considered to be progressive if the drug is withdrawn before much damage is done.

What causes pulmonary fibrosis?

In most types of pulmonary fibrosis, a specific cause cannot be found. The word 'idiopathic' means there is no known cause.

In a few cases, it's possible to identify a specific cause, such as:

- being exposed to certain types of dust – including wood or metal dusts or asbestos
- being exposed to allergens carried in the air – such as bird feathers or mould
- a viral or bacterial infection
- a side effect of a drug or treatment.

Some types of pulmonary fibrosis are more likely to occur when you have another condition such as rheumatoid arthritis or scleroderma. These are sometimes called connective tissue disease related ILDs or CT-ILD.

There are lots of different types of pulmonary fibrosis, with different causes. But often there is no known cause.

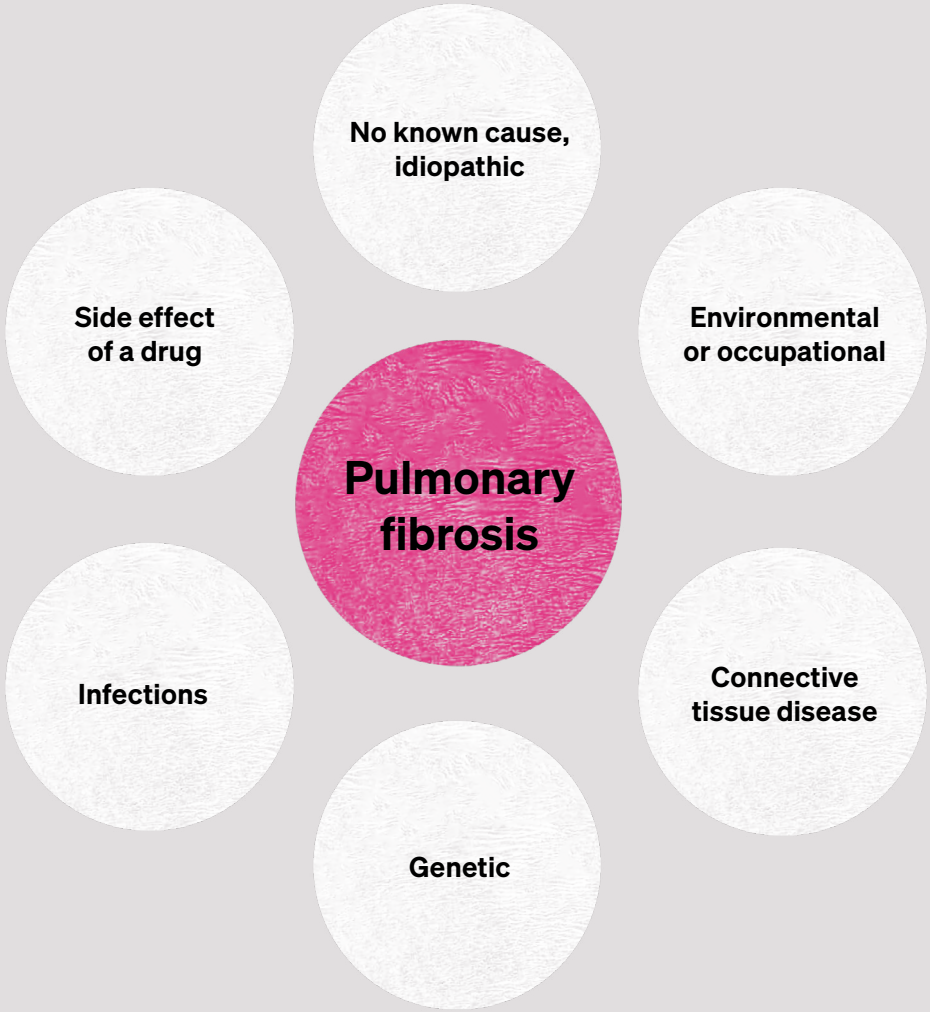
Is pulmonary fibrosis hereditary?

In a very few cases, two or more members of a family may develop pulmonary fibrosis. There are many complex ways in which it's possible to inherit a tendency to pulmonary fibrosis, and not all are well understood.

Current UK data sets suggest that less than 10% of people with pulmonary fibrosis have an inheritable form of the disease. Genetic testing isn't routinely available for people with pulmonary fibrosis. But there are some rare, but well-known, inheritable conditions that cause pulmonary fibrosis, such as dyskeratosis congenita.

If you, or a family member, has one then you may be offered genetic screening.

Smoking can cause some very specific kinds of pulmonary fibrosis such as desquamative interstitial pneumonia and respiratory bronchiolitis-interstitial lung disease.



What are the symptoms of pulmonary fibrosis?

The first symptom a lot of people notice is **getting out of breath** when they're exerting themselves, such as climbing a hill or stairs. But you might feel constantly short of breath, and not just when you're moving about.

Many forms of pulmonary fibrosis tend to occur after the age of 60, so some people wrongly think they're getting breathless because they're getting older. Don't assume this is the case – it may well get worse if it's not treated.

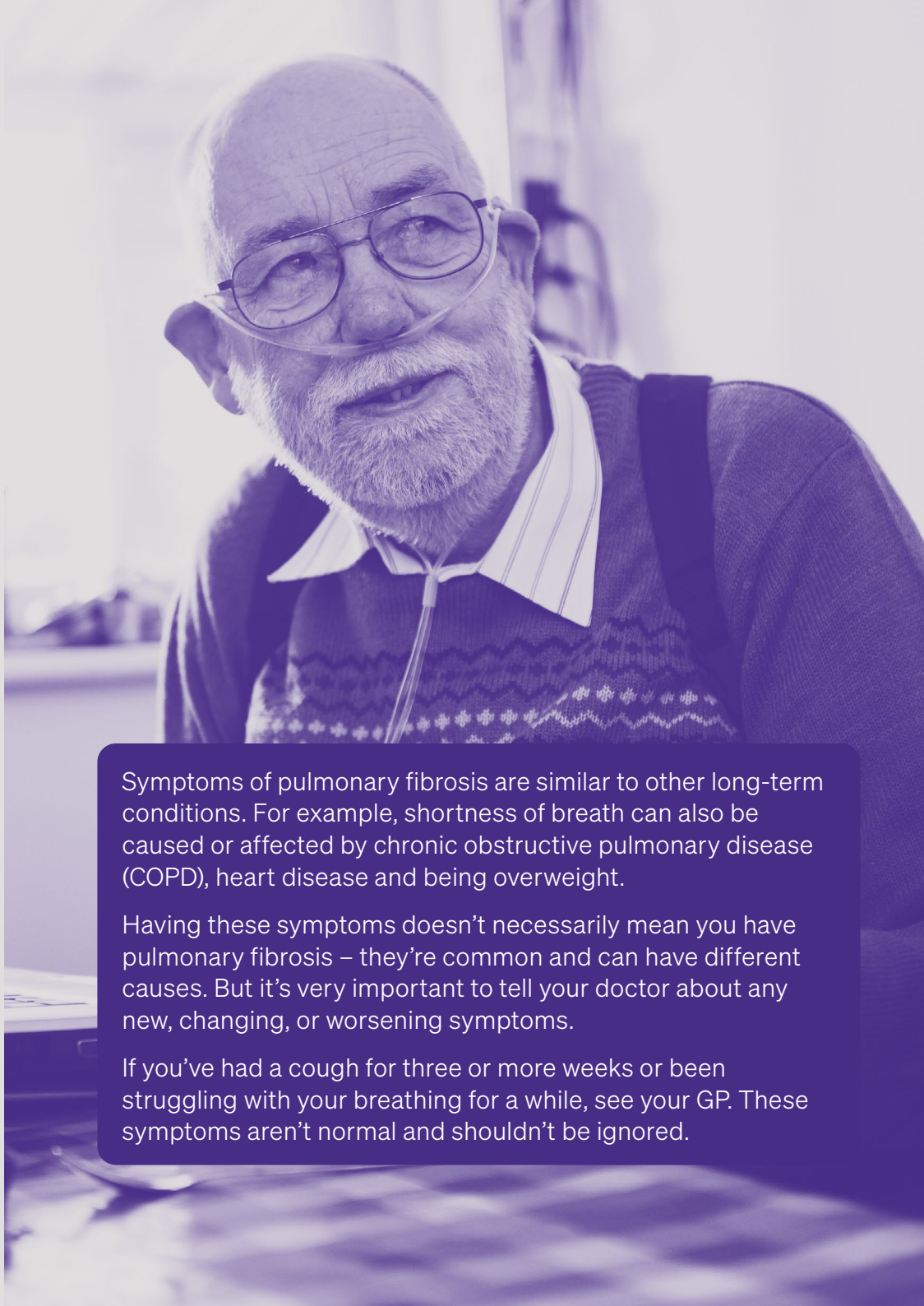
A cough that doesn't go away and feeling very tired all the time are two other symptoms of pulmonary fibrosis.

Some people with conditions associated with pulmonary fibrosis can lose weight or have a fever or aching or stiff joints and muscles. Others might have rashes, dry mouth and eyes, or poor circulation in fingers and toes (Raynaud's).

Another sign of pulmonary fibrosis is finger and toenail nail clubbing. You or your doctor may notice your nails changing shape to become like a drumstick: the tips of the fingers get bigger and the nails curve around the fingertips, and the base of the nail feels spongy.

Clubbing can also occur with other lung conditions, such as lung cancer, or as a result of heart or liver disease.

The different types of pulmonary fibrosis have similar symptoms. You'll have tests and speak to a specialist consultant to find out exactly which type you have.



Symptoms of pulmonary fibrosis are similar to other long-term conditions. For example, shortness of breath can also be caused or affected by chronic obstructive pulmonary disease (COPD), heart disease and being overweight.

Having these symptoms doesn't necessarily mean you have pulmonary fibrosis – they're common and can have different causes. But it's very important to tell your doctor about any new, changing, or worsening symptoms.

If you've had a cough for three or more weeks or been struggling with your breathing for a while, see your GP. These symptoms aren't normal and shouldn't be ignored.

How is pulmonary fibrosis diagnosed?

What should I do if I have symptoms of pulmonary fibrosis?

Make an appointment with your GP if you have symptoms that might be due to pulmonary fibrosis.

Your doctor will examine you, looking for other causes of breathlessness. They'll listen for crackles in your lungs and arrange a chest X-ray if they hear any.

If you have IPF, doctors listening to your chest can often hear crackles in your lungs that sound like opening velcro.

If there is any suspicion you may have pulmonary fibrosis, your doctor will refer you to a chest specialist at your local hospital. You may be referred on again for further investigation or specialist treatment to a specialist in pulmonary fibrosis and ILD if there isn't one in your local hospital.

What tests might I have to diagnose pulmonary fibrosis?

The hospital doctor will ask questions about your medical, family and work history and your symptoms. They will examine you, listen to your chest and assess the need for tests.

You may need tests such as:

- **a CT scan of your lungs**, which uses X-rays to produce detailed images of your lungs
- **breathing and lung function tests** to measure how well your lungs are working
- blood tests.

For some types of pulmonary fibrosis, the results from a CT scan can be very clear and allow a diagnosis to be made. For example, on a CT scan IPF often shows up as a distinctive pattern on the lungs. You might hear your doctor call this **honeycomb lung**. The image shows lots of empty pockets or bubbles appearing where more solid-looking lung tissue would normally appear.

Looking inside your lungs

Your doctor may need to look inside your lungs and possibly remove some cells or tissue for testing. Different procedures can be used to get some lung tissue:

- **flexible bronchoscopy** involves inserting a narrow tube through your nose or mouth, down into your lungs. You will have a local anaesthetic sprayed inside your nose and throat and often a sedative injection into your vein for your comfort. The tube has a camera on the end so the doctor can see inside your lungs. The doctor may flush some water through the tubing to remove and collect cells for analysis.
- **video-assisted thoracoscopy (VATS)** involves surgery under a general anaesthetic to get a larger piece of lung tissue. A surgeon makes keyhole incisions in your chest for a video-assisted surgical telescope to enter, and to remove tissue samples from your lungs. It's only undertaken if your specialist needs more tissue to make a diagnosis and treatment plan. Your doctor will discuss the risks with you. You'll stay in hospital for a few days for this test.
- **open lung biopsy** is a procedure undertaken by a specialist surgeon under general anaesthetic to take a sample from the lung.

Diagnosing pulmonary fibrosis is a joint effort by your specialist team. It will include doctors who are experts in lung conditions, surgery, X-rays and scans, and laboratory tests. The specialist nurse is an important part of this team and a good source of information and support for you. This multidisciplinary team (MDT) will review all your test results to reach a diagnosis.

Treatment for pulmonary fibrosis

Can pulmonary fibrosis be cured?

Once lung scarring occurs in the lungs it cannot be reversed. This means there is no cure for existing fibrosis, whatever the cause. But there are things you can do to control your symptoms and look after yourself.

How is pulmonary fibrosis treated?

Help to stop smoking

If you smoke, stopping is very important for your lung condition and overall health. Your GP can refer you to free help to stop smoking.

Drugs and medication

Drug treatments aim to stabilise or slow down the rate of scarring in the lungs. Specific drugs can be used to treat some people with pulmonary fibrosis who reach certain criteria:

- in people with IPF, Pirfenidone and Nintedanib can be used for those whose lung function tests fall within a certain range.
- in people with progressive fibrotic ILD (PF-ILD), Nintedanib may be used as well as or instead of immunosuppressant drugs. Forms of PF-ILD include systemic sclerosis associated ILD, autoimmune ILDs, and hypersensitivity pneumonitis.

Steroids or other immunosuppressant drugs are used to suppress inflammation in the lungs.

In conditions like hypersensitivity pneumonitis, it's key to avoid being exposed to the allergen causing symptoms, if the allergen can be identified.

Any drugs causing problems with your lungs will be stopped.

Lung transplant

For a very few people, having a lung transplant might be an option if the pulmonary fibrosis progresses and isn't stabilised by treatment. Transplants are rare – according to the NHS Annual Activity Report, 91 lung transplants were carried out in the UK in 2020-21, although this statistic was affected by the coronavirus pandemic. Not all of these were for people with pulmonary fibrosis.

Whether you can be considered for a lung transplant depends on factors that influence the chance of a successful outcome, such as your general health, other medical conditions and your weight. There is no age cut-off, but it's unusual to accept people much over 65 years old. There are not enough donor lungs available to meet demand. If your doctor thinks you might be suitable, you'll be referred to a transplant unit for further assessment and a decision.

There are significant risks in having a lung transplant. According to UK data, 81% of people are alive one year after a lung transplant, and 58% are alive five years afterwards. The survival rate after 10 years is 38%.

Best supportive care

Treating just the symptoms, rather than the cause of a disease, is often called **best supportive care**. As soon as you are diagnosed, your doctor, nurse or physiotherapist should talk to you about ways to reduce your symptoms and support your mental wellbeing. They may be able to do this at your usual place of care or you may be referred to a specialist in this area, usually a palliative care consultant.

Palliative care specialists are experts in symptom management and may help at various times during your illness, not just in the final stages. Palliative care focuses on controlling your symptoms, such as breathlessness, fatigue and anxiety. It may also stop treatments that are not working or causing side effects. The focus is on addressing issues to improve the quality of life for you, your family and carers.

Controlling your symptoms

For coughing, your doctor might treat problems that could be making it worse, such as heartburn (acid reflux) or a stuffy nose.

Tell your doctor if you have symptoms of acid reflux, such as heartburn, indigestion or a sour taste at the back of your mouth. There's evidence this may make inflammation and fibrosis worse. It can also make a cough worse.

Feeling out of breath can have a serious effect on your everyday life. It can be a frightening experience, too.

Pulmonary rehabilitation is an important way to help you cope with breathlessness by increasing your fitness and ability to cope with feeling out of breath. The course is designed to support and reassure you, as well as help your condition. You may find you can walk further, feel less breathless and generally feel more positive. Ask your team about this.

If your condition gets worse, the level of oxygen in your blood may fall and this may make you feel more breathless. If this happens, your doctor will refer you for an assessment for oxygen therapy. Using oxygen is a way of keeping more active, and later some people will use it at rest too. Your oxygen prescription will be tailored to meet your individual needs.

If you have distressing symptoms of breathlessness and really troublesome coughing, tell your doctor. They may prescribe you low doses of morphine and sedatives to help.

You may struggle to cope emotionally with having a serious condition like pulmonary fibrosis. You are not alone. Many people with a long-term lung condition feel anxious, have a low mood or symptoms of depression. It's important to look after your mental wellbeing. Your doctor or nurse will understand and can help.

Clinical trials

As well as existing therapies, you might be invited to take part in a medical study, also called a clinical trial, to investigate new treatments. It's not an option for everyone – if you want to know more, ask your doctor.

“I was amazed at how helpful pulmonary rehabilitation was. It told me things I needed to know. It gave me the confidence to go to the gym, even with my oxygen. I made a friend on the course and we went together.”

Lesley

Looking after yourself

If you have pulmonary fibrosis, you can do a lot to help yourself by leading a healthy lifestyle. Feeling very tired is a common symptom and health problems that used to be minor – such as catching a cold – can become more serious.

These tips will help you stay as healthy as possible:

- Have a flu jab each year and avoid being around people with colds. Talk to your doctor about the coronavirus vaccination to make sure you’re up to date.
- Ask for a pneumonia vaccination. This is a jab you have just once.
- Stay as fit as you can. There are many different ways to be active – find one that you enjoy. We have ideas at **blf.org.uk/keep-active**.
- Eat a healthy, balanced diet and maintain a healthy weight. Ask your doctor or nurse to refer you to a dietician, who can give you tailor-made advice, especially if you are losing weight. We also have advice at **blf.org.uk/eating-well**.
- Try techniques and positions to help your breathing. If you have pulmonary fibrosis, you may tend to breathe very fast and shallowly – a bit like panting. You can use techniques and positions to help you control and slow down your breathing. You can also use them to avoid getting too breathless when you exert yourself, and to help you recover when you do get out of breath. Try the different breathing techniques to find what helps you and practice the ones that help. Talk to your respiratory physiotherapist or nurse for help to find out what works for you.

Have a look at the techniques and positions online at **blf.org.uk/breathlessness**

“I adapt and stay positive.”

John was diagnosed with IPF four years ago.

I was invited to take part in a pulmonary rehabilitation study run by research physios and designed specifically for interstitial lung disease (ILD). It was a great help. I remain convinced exercise is a key way to help preserve my lung function.

I was first prescribed pirfenidone. I had a lot of side effects: nausea, vomiting, diarrhoea and a skin rash which required the regular application of SPF50 sun screen whenever I went out in the sun. I also felt very fatigued sometimes.

When test results indicated it wasn’t slowing the scarring as much as was hoped, I was switched to nintedanib. I was a bit nauseous initially, but I’ve found a good regime and manage my diet, which includes a good meal before taking the tablets.

I know the condition is progressing. I get more breathless when I do just normal everyday activities. I still work part-time, as this is important to me, but it is becoming more difficult. My recently acquired wheelable rucksack has made the one-hour commute a good bit easier. Little changes like these can make life easier.

I keep as active as I can. Badminton remains an important passion and I have many other hobbies and interests, I also exercise at home and do yoga.

I’ve started using ambulatory oxygen. I’d struggled with the concept, as I saw it as a sign my condition was progressing. But now I see it as a way of allowing me to continue to do the things I want to do.

My support group is vital

I’ve set up a support group for people with all types of ILD – and their families. I’m very aware that we don’t travel this road alone. Everyone has their own unique story about the impact of IPF on them.

Groups like ours can be a great way to get psychological support in a friendly and mutually supportive environment. It’s a way for people affected by IPF to take back some control and get support from others who are genuinely interested to hear how they are.

Getting support

Join a pulmonary fibrosis support group

Being diagnosed and living with a long-term lung condition can be challenging. Joining a pulmonary fibrosis support group can help you make new friends who understand what you're going through.

Find out more about pulmonary fibrosis support groups on our website at **blf.org.uk/support-for-you/pulmonary-fibrosis/support-groups**

No support group in your local area? Why not join HealthUnlocked, our online community at **blf.org.uk/support-for-you/web-community**. Chat to other people affected by lung disease, 24 hours a day, 7 days a week.

Other help and support for pulmonary fibrosis

www.actionpf.org – information and support for people diagnosed with pulmonary fibrosis, their families, friends and carers.

www.pulmonaryfibrosistrust.org – providing personal support for people living with pulmonary fibrosis.

www.versusarthritis.org – information about rheumatoid arthritis, connective tissue disease and immunosuppressant drugs.

Call our helpline on **0300 222 5800**

We are here to help:

- answer your questions, whether that's about coping with symptoms, your rights or finding equipment
- provide clear and trustworthy information about breathing problems and living with a lung condition
- you to get in touch with your local support group

Our friendly team are here Monday to Friday 9am to 5pm. Calling will cost the same as a local call. It's usually free, depending on your call package, even from a mobile.

Or visit **AsthmaAndLung.org.uk** to find support and information or to join our web community

Get support and information	**blf.org.uk/support-for-you**
Sign up to our newsletter	**blf.org.uk/signup**
Find your local support group	**blf.org.uk/breathe-easy**
join our web community	**healthunlocked.com/asthmalunguk-lung**

Did you find this information useful?

Scan the QR code with your phone's camera to donate today.

Each year over 400,000 people rely on our expert advice to make key decisions about their lung health. And today, you were one of them.

If you found this information useful and would like to help others access our expert advice, please consider making a kind gift. To donate: call **0300 222 5800**, visit **asthmaandlung.org.uk/leaflets** or scan the QR code. Thank you.

Helpline: **0300 222 5800**

AsthmaAndLung.org.uk

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